

Amyotrophic Lateral Sclerosis: First Case Report in Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran, Bandung

Ahmad Faried, Priandana Adya Eka Saputra, Alief Dhuha, Muhammad Zafrullah Arifin

Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran-Dr. Hasan Sadikin Hospital, Bandung

Abstract

Objective: Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that is incurable and results in paralysis of the muscles. Electromyography (EMG) is used to diagnosis amyotrophic lateral sclerosis. Although recently there is no cure for ALS, knowledge has increased significantly in the past several years and is helping those who are newly diagnosed.

Methods: This study reported for the first time in Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital, Bandung a 58-year old man who was presented in the institution with a history of weakness of both lower extremities for four months preceded by weakness of both upper extremities since the previous month. There was no history of any medical illness or any chronic medication. The patient then underwent EMG studies, followed by muscle biopsy.

Results: Electromyography and histopathological results confirmed a diagnosis of ALS.

Conclusions: This case was so exceptional, since ALS occurrence in Neurosurgery Centre is extremely rare, and the diagnosis can only be established through EMG and histopathological of muscle biopsy studies.

Received:
April 24, 2018

Revised:
December 20, 2018

Accepted:
March 19, 2019

Keywords: The fist reported of ALS in Neurosurgery Bandung, electromyography studies

pISSN: 2302-1381; eISSN: 2338-4506; <http://doi.org/10.15850/ijhs.v7n1.1237>
IJHS. 2019;7(1):56-60

Introduction

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease characterized by progressive muscular paralysis reflecting degeneration of motor neurons (MNs) in the primary motor cortex, brainstem and spinal cord. Amyotrophy refers to the atrophy of muscle fibers, leading to weakness of affected muscles and fasciculations. Lateral Sclerosis refers to hardening of the anterior and lateral corticospinal tracts as MNs in these areas degenerate and are replaced by gliosis; hence the electromyography (EMG) and the histopathological examination should be performed in order to confirm the diagnosis.¹

Correspondence:

Ahmad Faried, Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran-Dr. Hasan Sadikin Hospital, Bandung
Jl. Pasteur No. 38, Bandung, Indonesia
e-mail: faried.fkup@gmail.com

The etiology of ALS has not been well understood; but it seems that various mechanisms leading to ALS been propose, such as (i) gene mutation, various genetic mutations can lead to inherited ALS which appears nearly identical to the noninherited form, (ii) chemical imbalance people with ALS generally have higher than normal levels of glutamate, a chemical messenger in the brain, around the nerve cells in their spinal fluid. High concentration of glutamate is known to be toxic to some nerve cells, (iii) disorganized immune response, sometimes a person's immune system begins attacking some of his or her body's own normal cells, which may lead to the death of nerve cells.²⁻⁴ International data showed the incidence of ALS is 1.9 patients per 100,000 people in the population and ALS prevalence is 6 per 100,000 people in the population; The highest incidence of ALS found in the population aged over 50 years.

Table 1 Needle EMG Result that Supported Final Diagnosis of Amyotrophic Lateral Sclerosis

Muscles	Spontaneous Activity				MUAP's			Recruitment Pattern	IP	
	Fibs	PSW	Fasc	Ltc	Amp (mV)	Dur. (min)	Poly.P			
Right Muscle										
M. Gastrocnemius	↑	+	+	-	-	0.1-0.3	2-6	+	↓	↓
M. Tibialis anterior	↑	+	+	-	-	0.1-1.0	2-10	+	↓	↓
M. Vastus lateralis	↑	+	+	-	-	0.1-1.0	2-14	+	↓	↓
M. Gluteus maximus	↑	+	+	-	-	0.1-2.0	2-18	+	↓	↓
M. Deltoid	N	-	-	-	-	0.1-1.6	2-8	-	N	N
Left Muscle										
M. Gastrocnemius	↑	+	+	-	-	0.1-2.0	2-14	+	↓	N

Notes:

Amp=Amplitude, Dur=Duration, Fasc=Fasciculations, Fib=Fibrillation potentials, IA=Insertional activity, IP=Interference Pattern, Ltc=Latency, MUAP=Motor unit action potential, PSW: Positive sharp wave, Poly.P=Polyphasic potential,

Assuming that 15 % of Indonesia's population aged over 50 years (total population in 2015 are 260 million people); the prevalence of ALS in Indonesia in 2015 is estimated at 2,500 people.⁵ However, the incidence nor observation never been reported from Indonesia, except one report from 2001-2012 survey results regarding ALS and Parkinson in Papua, Indonesia.⁶

Herewith, the study reported for the first time at Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran-Dr. Hasan Sadikin General Hospital, Bandung, a case of 58-years old man with weakness on his four extremities, thus diagnosed as ALS according to the histopathological and EMG findings. Although, the study might not represent the general population in Indonesia and requires further investigation.

Case

A 58-year old man came to our center with weakness of both lower extremities for 4 months as the chief complaint; this was also accompanied with weakness of both upper extremities since the previous month (Fig. 1a-c). The complaint was initially felt four months prior to consultation; at the moment the patient was unable to stand on his toes and

tiptoed. The complaint further progressed, with the right side is weaker than the left one. At the time of consultation, the patient was already unable to stand and walk properly. There were no histories of any medical illness or any chronic medication. The patient had already undergone hospitalization by our fellow neurologist who further consulted the patient to our department.

At presentation, the patient was fully conscious and in moderate-ill condition. Vital signs were within normal limit. On both lower extremities there were atrophies with decreased power (3/5). There were no sensory, proprioceptive or autonomic deficits. Any other physical examination was within normal limit. Neuroimaging modalities such as head and complete spinal MRI had failed to show any pathology lesion. Nerve conduction study shows decreased compound muscle action potential (CMAP) on bilateral peroneal and tibial nerve with intact sensory function. Needle electromyography shows increased insertion activities, complex repetitive discharges, fibrillation, positive sharp waves, polyphasic, decreased recruitment on bilateral gastrocnemius muscles suggestive for myogenic lesion. An initial diagnosis of motor neuropathy disease (MND) was made (Table 1 and 2).

Muscle biopsy was then performed to this

patient. Muscle biopsy is needed only rarely but may be considered if the presentation of ALS is atypical. The results will confirm the presence of signs of denervation and re-innervation or may lead to an alternative diagnosis. The presence of small, angular fibers is consistent with neurogenic atrophy (denervation). Fiber-type grouping is consistent with re-innervation. The biopsy was performed at the right gastrocnemius muscle under local anesthesia. A muscle tissue sample, sized about 1 cm³ was sent to a pathologist at Dr. Hasan Sadikin General Hospital, Bandung. This muscle biopsy shows neurogenic atrophic changes, with clumps of pyknotic nuclei, large group (fascicular) atrophy, hypertrophic fibers, and scattered angulated atrophic fibers (Hematoxylin and eosin or HE stain, ×100). Markedly atrophic muscle obtained at biopsy shows end-stage myopathic changes with diffuse fatty infiltration (HE stain, ×200). The final diagnosis for our patient was ALS. The patient was discharged, on his own will, with

slight balance disturbance and no post-biopsy complication was observed. The patient died one month later of cardiorespiratory failure, seven months after symptom onset.

Discussion

Amyotrophic lateral sclerosis is a neurodegenerative disease that paralyzes the muscles because of the loss of functioning motor neurons. Amyotrophy refers to the atrophy of the muscle fibers, which then produces spasms and muscle weakness. Lateral sclerosis refers to the hardening of the anterior and lateral horn of the corticospinal tract.⁷ There are many forms of ALS which include: a) progressive bulbar palsy; b) progressive muscular atrophy; c) primary lateral sclerosis; d) flail arm syndrome; e) flail leg syndrome.⁷ Each form of ALS has its own set of symptoms but all somehow will lead to the same degenerative and eventually the

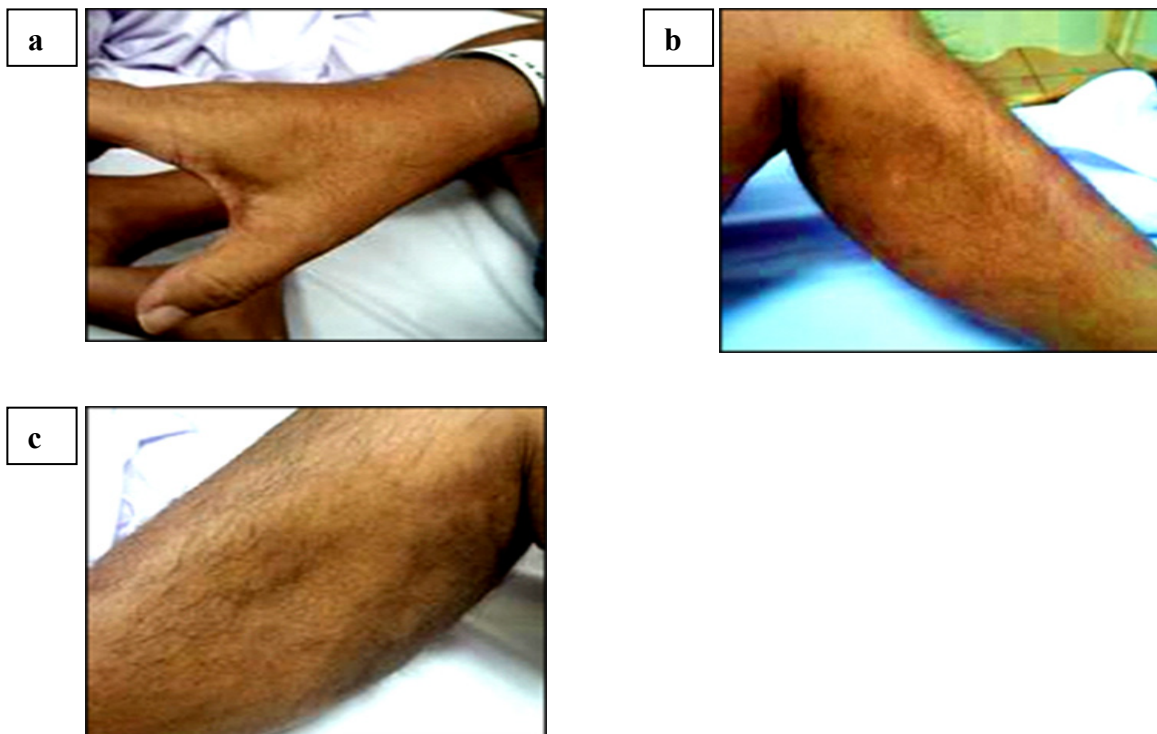


Fig. 1 Clinical Inspection Reveals Symmetrical Atrophies on Both Hands and Legs. Muscle Atrophy at Right Hand (a); Muscle Atrophy at Left Leg (b); Muscle Atrophy at Right Leg (c)

Table 2 Needle EMG Result that Supported Final Diagnosis of Amyotrophic Lateral Sclerosis (Continued from Table 1)

Muscles	IA	Spontaneous Activity				MUAP's			
		Fibs	PSW	Fasc	Amp (mV)	Poly.P	Config.	Recruitment	
First Dorsal Interosei (R)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	
Brachioradialis (R)	Normal	None	1+	None	0.2-1.0	None	Normal	Normal	
Bicep Brachii (R)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	
Deltoid (R)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	
Tricep Brachii (L)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	
Tibialis Anterior (R)	Increased	1+	1+	None	0.2-1.0	None	Normal	Reduced	
Peroneus Longus (R)	CRD	2+	2+	None	0.2-1.0	None	Normal	Reduced	
Gastrocnemius (R)	Increased	1+	None	None	0.1-0.3	Few	Normal	Reduced	
Vastus Medialis (R)	Normal	1+	None	None	0.2-1.0	None	Normal	Reduced	
Tibialis Anterior (L)	Normal	1+	1+	None	0.2-1.0	Few	Normal	Reduced	
Gastrocnemius (L)	Normal	1+	1+	None	0.1-0.3	Few	Normal	Reduced	
First Dorsal Interosei (L)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	
Deltoid (L)	Normal	None	None	None	0.2-1.0	None	Normal	Normal	

Notes:

Amp=Amplitude, Dur=Duration, Fasc=Fasciculations, Fib=Fibrillation potentials, IA=Insertional activity, IP=Interference Pattern, Ltc=Latency, MUAP=Motor unit action potential, PSW: Positive sharp wave, Poly.P=Polyphasic potential, IP: Interference Pattern

worst outcome. Amyotrophic lateral sclerosis is terminal and eventually leads to death caused by respiratory distress. The aim was to explain the different types of ALS, how imaging is currently being used, sometimes it failed, to diagnose the disease and a few palliative care methods used to care for ALS patients.

The progressive muscular atrophy (PMA) is one of the type of ALS; it is characterized by degeneration of the lower motor neurons without any or less symptoms of upper motor neuron degeneration. These patients lose control of their limbs and trunk. The PMA progresses slowly with bulbar and respiratory problems appearing later.⁸ The PMA eventually leads to mainstream ALS which will slow the patient down and lead to respiratory problems.

Magnetic resonance imaging (MRI) is the only imaging modality that is currently used to diagnose ALS. The majority of the time MRI is used to rule out other ALS mimicking diseases. In the effort to gain knowledge about the ALS disease new MRI techniques have been developed to image the brain

differently. Somehow not in all case it would be useful. Other modalities to diagnose ALS are EMG. Often used to rule out other muscular disorders, EMG measures the amount of muscle movement over a particular area, then it is shown in wave form.⁹ General feature of MND histopathology: a) decreased of large motor neuron with focal astroglyosis; b) senescent changes; c) intra cytoplasmic inclusion; d) proximal and distal axonopathies distal with axonal spheroid; e) tractus degeneration; f) motoric fibers degeneration, motor end-plates and muscle atrophies.¹⁰

Even though ALS has been around since the 1800s it was not a very well known disease until Lou Gehrig was diagnosed in the 1940s.³ There is a lot that is unknown about ALS and what causes it. Although there is no cure for ALS, knowledge has increased significantly in the past several years and is helping those who are newly diagnosed. By keeping the patient nourished and slowing down the progression of the disease, the efforts could help to sustain life. The cellular therapy

with stem cells or induced pluripotent stem cells (iPSCs), surrounded by a deal of great technical challenge, is presented with a future perspective of neuro-regeneration as well as of neuro-protection of motor-neurons in ALS.

In conclusion, ALS is particularly rare at

Neurosurgery Centre and the diagnosis along with the treatment is difficult to be established. The imaging modalities alone could be challenging the diagnosis of ALS. In this study, the diagnosis of ALS can only be confirmed by means of EMG and histopathological studies.

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